Vascular cases

Case 1 (Master 11/2006)

A middle-aged woman suffering from painful plum colored edematous nodules & plaques on the limbs, neck & face together with fever, malaise, arthralgia & conjunctivitis. There was a preceding upper respiratory infection.

Lab investigations: elevated ESR, neutrophilia

a) What is your diagnosis? How would you confirm it?

b) Give an account on DD & treatment?

c) Comment on pathogenesis & mention other associations?

Answer:

a) Sweet's syndrome

Key points:

- Middle-aged woman (female: male, 3.5:1)
- Painful **plum** colored
- Fever
- Upper respiratory infection
- Neutrophilia

Histopathology:

Upper & mid-dermal infiltration of neutrophils, the infiltrate less often includes lymphocytes, histiocytes & eosinophils. Leukocytoclasia is a prominent feature, without evidence of vasculitis. Dermal edema & RBCs extravasation may be present.

b) DD:

- Acne fulminans
- Acral erythema
- Behcet's disease
- Cellulitis
- Cutaneous small vessels vasculitis
- Deep fungal infection
- Erythema elevatum diutinum

- Erythema multiforme
- Erythema nodosum
- Granuloma annulare
- Granuloma faciale
- Sarcoidosis
- Syphilis
- Urticarial vasculitis

Treatment:

- Prednisone 40-6- mg/day (0.5-2.5 mg/kg/day), tapering over 4-6 weeks.
- Others: cyclosporine (5-10 mg/day), colchicines (1.5 mg/day), isotretinoin, methotrexate, dapsone (100-200 mg/day)

c) Pathogenesis:

- Ig & complement-mediated activation & mobilication of neutrophils.
- Antigen or superantigen-induced T-cell dependent cellular immune reaction.
- Local & systemic dysregulation of cytokine secretion (IL-1, G-CSF, GM-CSF, INF- δ)
- Associated with HLA-BW 54
- Altered function of neutrophils.

Other associations:

- Malignant: lymphoma, breast, ovarian, vaginal, rectal, prostate, testicular & endometrial cancers
- Inflammatory: ulcerative colitis, crohn's disease, rheumatoid arthritis, Behcet's syndrome
- Infections: staphylococcus, streptococcus, TB, meningitis, hepatitis, HIV
- Others: drugs, pregnancy, renal stones, immunization, photoinduction.

Case 2 (Master 11/2012)

A 28-years old female patient presented with edematous painful erythematous papules & plaques on the lower eyelids & cheeks. Some of the papules look like vesicles but are firm upon palpation. The eruption occurred after upper respiratory tract infection. The patient had fever & leukocytosis. Skin biopsy showed massive infiltrate of neutrophils in the upper dermis. Leucocytoclasia is frequently present.

a) What is the most likely diagnosis?

b) What are the DD?

c) What is the treatment of the disease?

Answer:

a) Sweet's syndrome

Key points:

- 28-years old female (Middle-aged, female: male, 3.5:1)
- Papules look like vesicles but are firm (pseudovesicle)
- Upper respiratory tract infection
- Fever & leukocytosis
- Neutrophils in the upper dermis

b) DD: see before

c) *Treatment:*

- Prednisone 40-6- mg/day (0.5-2.5 mg/kg/day), tapering over 4-6 weeks.
- Others: cyclosporine (5-10 mg/day), colchicines (1.5 mg/day), isotretinoin, methotrexate, dapsone (100-200 mg/day)

Case 3 (Diploma 11/2010)

A female patient 40 years old presented with acute tender erythemetous nodules over the pretibial areas bilaterally & symmetrically distributed. The patient also complained of arthralgia & fever.

a) What is the provisional diagnosis & pathogenesis of this disease?

b) What are the possible most common causes?

c) How can you manage this condition?

Answer:

a) Erythema nodosum

Key points:

- Female patient 40 years old (20-40 years, female: male, 3:1)
- Tender erythemetous nodules
- Pretibial areas bilaterally & symmetrically
- Fever

Pathogenesis:

It is considered a reactive erythema due to many etiological factors. It may be due to formation of immune complexes & their deposition in the vessels in deep dermis & adipose tissue (septal panniculitis)

b) The most common causes: streptococcal infections, drugs, sarcoidosis, ulcerative colitis.

c) Management:

Histopathological examination:

Deep skin biopsy, (septal panniculitis without vasculitis): septal inflammatory infiltrate associated with septal vascular endothelial swelling, edema & hemorrhage. There is predominance of lymphocytes, histiocytes & giant cells. No leukocytoclastic vasculitis or fat necrosis.

Miescher's radial granulomas is consisting of small, well-defined, nodular aggregations of small histiocytes around a central stellate cleft.

Laboratory investigations:

- ↑ ESR, mild anemia, leukocytosis
- Chest X-ray: hilar adenopathy or infiltrates (sarcoid Lofgren syndrome)
- ↑ASO-titre, throat swab & culture (streptococcal)
- Intradermal skin tests (TB, sarcoidosis, deep mycosis)

Treatment:

- Treatment of underlying cause
- Complete bed rest
- NSAID (indomethacin)
- Potassium iodide (900mg/day 3-4 weeks)

Case 4 (Diploma 4/2016)

A 35 years old female presented with bilateral red tender poorly defined nodules 2-5 cm in diameter on the anterior legs. The patient has experienced mild arthralgia few days preceding the rash.

a) What is the most likely diagnosis?

b) What are the DD?

c) What is the management?

Answer:

a) Erythema nodosum

Key points:

- 35 years old female (20-40 years, female: male, 3:1)
- Bilateral red tender poorly defined nodules (felt better than seen)
- Anterior legs

b) DD:

- Erythema induratum of Bazin
- Erythema nodosum leprosum
- Polyarteritis nodosa

c) Management: see before

Case 5 :

A 38 years old female presented with tender inflammatory nodules on chin of tibia, the patient complained also of fever & arthralgia of ankle joint.

a) What is your diagnosis?

b) What are the DD?

c) What is the management?

Answer:

a) Erythema nodosum

Key points:

- 38 years old female
- Tender inflammatory nodules on chin of tibia
- Fever & arthralgia

b) see before

c) see before

Case 6 (Master 11/2013)

A 42 years old female complaining from persistence painful rather than pruritic wheal-like lesions distributed all over the body. Lesions leave post-inflammatory hyperpigmentation.

a) What is your diagnosis?

b) What are the associated internal diseases?

c) What is the available treatment for this case?

Answer:

a) Urticarial vasculitis syndrome

Key points:

- 42 years old female (30-50 y, female: male, 3:1)
- Painful rather than pruritic
- Post-inflammatory hyperpigmentation

b) Arthralgia, abdominal pain, uveitis, glomerulonephritis

c) Treatment:

1st line: H1 &H2 antihistamines, NSAID, dapsone, corticosteroid

2nd line: colchicin, hydroxychloroquine, azathioprine, methotrexate

3rd line: mycophenolate mofetil, IVIg, cyclosporine, plasmapheresis

Case 7:

A 56 years old male suffering from renal failure presented with livedo reticularis, ecchymosis & necrotic black eashars on abdomen & thigh.

a) What is your diagnosis?

b) Mention other cutaneous manifestations of renal failure?

Answer:

a) Polyarteritis nodosa

Key points:

- Renal failure
- Livedo reticularis
- Ecchymosis
- Necrotic black eashars

b) see next paper

Renal

A) Related to multisystem disorder: (5 S, vessel, F N)

- **S**LE - **S**arcoidosis - **S**cleroderma - 1ry **S**ystemic amyloidosis - Tuberous **S**clerosis

- Vasculitis: HSP, PAN, LCV - Fabry's - Neurofibromatosis

B) Related to end-stage renal disease:

** Signs & symptoms:* جلده ينشف و لونه يتغير و يجي عليه فطريات

1- Pruritus

2- Xerosis, acquired ichthyosis

3- Keratotic pits on palm & sole

4- Color changes: pallor, yellow, ecchymosis, hyperpigmentation, uremic frost

5- Onychomychosis & tinea pedis

6- Nail: Muchreke nails, half & half nail, pale nail, splinter he

** Specific disorders:*

1- Perforating disorders: Kyrle's disease

2- Metastatic calcification

3- Bullous diseases: PCT, pseudoporphyria

C) Related to dialysis: 4 P - HANG

1- **P**ruritus

2- Acquired **p**erforating dermatosis

3- **B**ullous dermatosis

4- **P**seudo-PCT

5- Splinter **H**ge

6- **A**cne

7- Uremic **n**europathy

8- **G**ynecomastia

D) Related to transplantation: Increased incidence of infections, cancer, cushing syndrome

Case 8:

Child patient 8 years old complaining of palpable purpuric lesions on lower extremeties, these lesions are preceded by upper respiratory tract infection. There are also headache, anorexia, fever, acute abdominal pain & acute arthralgia.

a) What is your diagnosis?

b) What is the management?

Answer:

a) Henoch-Schonelin purpura

Key points:

- Child patient 8 years old (2-11 years)
- Palpable purpuric lesions
- Lower extremeties
- Preceded by upper respiratory tract infection
- Acute abdominal pain & acute arthralgia

b) Management:

Histopathological examination: granulocytes in the walls of arterioles or venules.

DIF: IgA, C3 & fibrinogen within vessels walls

Treatment:

1st line: supportive care, bed rest, NSAIDs

2nd line: dapsone & colchicines, systemic corticosteroid for arthritis & abdominal pain, azathioprine

3rd line: mycophenolate mofetil, corticosteroid + tacrolimus, IVIg, plasmapheresis

Case 9:

A 52 years old female, developed a rash on her hand, feet & extensors. She was unwell for a few days before with a cold. On examination, there are numerous circular lesions approximately 2 cm in diameter which have a purpuric or pale center. She states, new lesions develop whenever there is any trauma to site.

a) What is your diagnosis?

b) What are the causes?

c) What is the treatment?

Answer:

a) Erythema multiforme

Key points:

- Hand
- Preceded by cold
- Circular lesions purpuric or pale center (target lesion)
- New lesions develop after trauma (Koebner phenomenon)

b) Causes:

- Infections: **HSV (most common cause)**, AIDS, HBV
- Drugs: sulfonamide, contraceptive pills
- Autoimmune & vascular disease: LE, DM, PAN, Wegener's granulomatosis
- Malignancies: carcinoma, leukemia
- Pregnancy
- X-ray
- Sarcoidosis
- Idiopathic

c) Treatment:

1- Prophylactic: HSV-associated EM, prophylaxis for 6 months

- Oral acyclovir (10mg/kg/d in divided doses), or
- Valacyclovir (500-100mg/d), or
- Famciclovir (250mg twice daily)

If resistant to antiviral:

- Azathioprine (100mg/d)
- Prednisone (0.5mg/d)
- Dapsone
- Cyclosporine
- PUVA

2- Treatment of acute attacks:

- Treatment of underlying cause
- Systemic Ab to guard against 2ry infection
- Minor cases: symptomatic ttt
- Major cases: prednisone (0.5-1 mg/kg/d)